

**WEST**[Help](#)[Logout](#)[Interrupt](#)[Main Menu](#)[Search Form](#)[Posting Counts](#)[Show 8 Numbers](#)[Edit 8 Numbers](#)[Preferences](#)**Search Results -**

Term	Documents
(3 AND 1).USPT,JPAB,EPAB,DWPI,TDBD.	18

**Database:**

US Patents Full-Text Database  
 JPO Abstracts Database  
 EPO Abstracts Database  
 Derwent World Patents Index  
 IBM Technical Disclosure Bulletins

L1 and L3

**Refine Search:****Clear****Search History****Today's Date: 8/17/2000**

<u>DB Name</u>	<u>Query</u>	<u>Hit Count</u>	<u>Set Name</u>
USPT,JPAB,EPAB,DWPI,TDBD	L1 and L3	18	<u>L8</u>
USPT,JPAB,EPAB,DWPI,TDBD	L5 and L3	0	<u>L7</u>
USPT,JPAB,EPAB,DWPI,TDBD	L5 and L2	0	<u>L6</u>
USPT,JPAB,EPAB,DWPI,TDBD	L1 and L4	4	<u>L5</u>
USPT,JPAB,EPAB,DWPI,TDBD	(knockout mutation)	45	<u>L4</u>
USPT,JPAB,EPAB,DWPI,TDBD	(spinal muscular) adj (atrophy)	194	<u>L3</u>
USPT,JPAB,EPAB,DWPI,TDBD	(Smn gene)	9	<u>L2</u>
USPT,JPAB,EPAB,DWPI,TDBD	(transgenic mouse)	2613	<u>L1</u>

**WEST**[Generate Collection](#)

Search Results - Record(s) 1 through 9 of 9 returned.

☒ 1. Document ID: US 6080577 A

L2: Entry 1 of 9

File: USPT

Jun 27, 2000

US-PAT-NO: 6080577

DOCUMENT-IDENTIFIER: US 6080577 A

TITLE: Survival motor neuron (SMN) gene: a gene for spinal muscular atrophy

Full	Title	Citation	Front	Review	Classification	Date	Reference	Claims	KWIC	Draw Desc	Image
------	-------	----------	-------	--------	----------------	------	-----------	--------	------	-----------	-------

☐ 2. Document ID: US 5935783 A

L2: Entry 2 of 9

File: USPT

Aug 10, 1999

US-PAT-NO: 5935783

DOCUMENT-IDENTIFIER: US 5935783 A

TITLE: Genes mapping in the digeorge and velocardiofacial syndrome minimal critical region

Full	Title	Citation	Front	Review	Classification	Date	Reference	Claims	KWIC	Draw Desc	Image
------	-------	----------	-------	--------	----------------	------	-----------	--------	------	-----------	-------

☐ 3. Document ID: US 5882868 A

L2: Entry 3 of 9

File: USPT

Mar 16, 1999

US-PAT-NO: 5882868

DOCUMENT-IDENTIFIER: US 5882868 A

TITLE: Method of diagnosing spinal muscular atrophy

Full	Title	Citation	Front	Review	Classification	Date	Reference	Claims	KWIC	Draw Desc	Image
------	-------	----------	-------	--------	----------------	------	-----------	--------	------	-----------	-------

☐ 4. Document ID: JP 08228785 A

L2: Entry 4 of 9

File: JPAB

Sep 10, 1996

PUB-NO: JP408228785A  
DOCUMENT-IDENTIFIER: JP 08228785 A  
TITLE: SURVIVAL MOTOR NEURON (SMN) GENE: SPIRAL MUSCULAR  
ATROPHY

Full	Title	Citation	Front	Review	Classification	Date	Reference	Claims	RWC	Draw Desc	Image
------	-------	----------	-------	--------	----------------	------	-----------	--------	-----	-----------	-------

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☐ 5. Document ID: EP 711833 A2

L2: Entry 5 of 9                      File: EPAB                      May 15, 1996

PUB-NO: EP000711833A2  
DOCUMENT-IDENTIFIER: EP 711833 A2  
TITLE: Survival motor neuron (SMN) gene: a gene for spinal  
muscular atrophy

Full	Title	Citation	Front	Review	Classification	Date	Reference	Claims	RWC	Draw Desc	Image
------	-------	----------	-------	--------	----------------	------	-----------	--------	-----	-----------	-------

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☐ 6. Document ID: EP 708178 A1

L2: Entry 6 of 9                      File: EPAB                      Apr 24, 1996

PUB-NO: EP000708178A1  
DOCUMENT-IDENTIFIER: EP 708178 A1  
TITLE: Survival motor neuron (SMN) gene: a gene for spinal  
muscular atrophy

Full	Title	Citation	Front	Review	Classification	Date	Reference	Claims	RWC	Draw Desc	Image
------	-------	----------	-------	--------	----------------	------	-----------	--------	-----	-----------	-------

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☒ 7. Document ID: AU 9964696 A, WO 200021553 A2, EP 999270 A1

L2: Entry 7 of 9                      File: DWPI                      May 1, 2000

DERWENT-ACC-NO: 2000-317843  
DERWENT-WEEK: 200036  
COPYRIGHT 2000 DERWENT INFORMATION LTD

TITLE: Use of interferons for increasing interferon regulatory  
factor or stat factor to regulate the survival motor neuron  
gene expression, and treating spinal muscular atrophy

Full	Title	Citation	Front	Review	Classification	Date	Reference	Claims	RWC	Draw Desc	Image
------	-------	----------	-------	--------	----------------	------	-----------	--------	-----	-----------	-------

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☐ 8. Document ID: EP 711833 A2

L2: Entry 8 of 9

File: DWPI

May 15, 1996

DERWENT-ACC-NO: 1996-232098

DERWENT-WEEK: 199624

COPYRIGHT 2000 DERWENT INFORMATION LTD

TITLE: Human survival motor neuron gene T-BCD541, variant C-BCD541 and murine equiv. - useful to develop primers and probes for in vitro detection of motor neuron diseases e.g. spinal muscular atrophy

Full	Title	Citation	Front	Review	Classification	Date	Reference	Claims	KWIC	Draw Desc	Image
------	-------	----------	-------	--------	----------------	------	-----------	--------	------	-----------	-------

☒ 9. Document ID: US 6080577 A, EP 708178 A1, AU 9534369 A, CA 2160937 A, JP 08228785 A, EP 711833 A3, AU 702252 B

L2: Entry 9 of 9

File: DWPI

Jun 27, 2000

DERWENT-ACC-NO: 1996-202055

DERWENT-WEEK: 200036

COPYRIGHT 2000 DERWENT INFORMATION LTD

TITLE: New isolated survival motor neuron gene - used to develop prods. for the diagnosis and treatment of motor neuron diseases

Full	Title	Citation	Front	Review	Classification	Date	Reference	Claims	KWIC	Draw Desc	Clip Img	Image
------	-------	----------	-------	--------	----------------	------	-----------	--------	------	-----------	----------	-------

[Generate Collection](#)

Term	Documents
SMN.DWPI,TDBD,EPAB,JPAB,USPT.	226
SMNS	0
GENE.DWPI,TDBD,EPAB,JPAB,USPT.	120974
GENES.DWPI,TDBD,EPAB,JPAB,USPT.	36178
(SMN ADJ GENE).USPT,JPAB,EPAB,DWPI,TDBD.	9

[Display](#)

10

Documents, starting with Document:

9

**Display Format:**

TI

[Change Format](#)

### Status: Path 1 of [Dialog Information Services via Modem]

### Status: Initializing TCP/IP using (UseTelnetProto 1 ServiceID pto-dialog)  
Trying 3106900061...Open

DIALOG INFORMATION SERVICES

PLEASE LOGON:

\*\*\*\*\* HHHHHHHH SSSSSSSS?

### Status: Signing onto Dialog

\*\*\*\*\*

ENTER PASSWORD:

\*\*\*\*\* HHHHHHHH SSSSSSSS? \*\*\*\*\*

Welcome to DIALOG

### Status: Connected

Dialog level 00.07.20D

Last logoff: 15aug00 10:23:15

Logon file001 17aug00 10:04:59

\*\*\* ANNOUNCEMENT \*\*\*

NEW FILE RELEASED

\*\*\*Prous Science Daily Essentials (Files 458, 459)

\*\*\*WIPO/PCT Patents Fulltext (File 349)

UPDATING RESUMED

\*\*\*Datamonitor Market Research (File 761)

\*\*\*Dissertation Abstracts Online (File 35)

\*\*\*GPO Monthly Catalog (File 66)

\*\*\*Bridge World Markets News (File 609,809)

\*\*\*Fort Worth Star-Telegram (File 427)

\*\*\*

RELOADED

\*\*\*Canadian Business Directory (File 533)

\*\*\*D&B International Dun's Market Identifiers (File 518)

\*\*\*D&B European Dun's Market Identifiers (File 521)

\*\*\*Kompas Canada (File 594)

\*\*\*CANCERLIT (File 159)

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OneSearch simply BEGIN FIRST for coverage from Dialog's  
broad spectrum of news wires.

>>> Enter BEGIN HOMEBASE for Dialog Announcements <<<

>>> of new databases, price changes, etc. <<<

\*\*\*\*

KWIC is set to 50.

HIGHLIGHT set on as '\*'

File 1:ERIC 1966-2000/Jul 26

(c) format only 2000 The Dialog Corporation

Set	Items	Description
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?b 155,5,73

17aug00 10:05:15 User259876 Session D101.1

\$0.42 0.120 DialUnits File1

\$0.42 Estimated cost File1

\$0.01 TYMNET

\$0.43 Estimated cost this search

\$0.43 Estimated total session cost 0.120 DialUnits

SYSTEM:OS - DIALOG OneSearch

File 155:MEDLINE(R) 1966-2000/Oct W1

(c) format only 2000 Dialog Corporation

File 5:Biosis Previews(R) 1969-2000/Aug W3

(c) 2000 BIOSIS

File 73:EMBASE 1974-2000/Jul W4

(c) 2000 Elsevier Science B.V.

**\*File 73: Update codes are currently undergoing readjustment.**

For details type Help News73.

Set	Items	Description
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-----	-------	-------

?s (SMN (w) gene?)

Processing

991	SMN
-----	-----

5347215	GENE?
---------	-------

S1	409	(SMN (W) GENE?)
----	-----	-----------------

?s (spinal (w) muscular (w) atrophy)

348407	SPINAL
--------	--------

267218	MUSCULAR
--------	----------

109072	ATROPHY
--------	---------

S2	4026	(SPINAL (W) MUSCULAR (W) ATROPHY)
----	------	-----------------------------------

?s (transgenic (w) (mouse or mice))

76050	TRANSGENIC
-------	------------

1181644	MOUSE
---------	-------

1071999	MICE
---------	------

S3	40099	(TRANSGENIC (W) (MOUSE OR MICE))
----	-------	----------------------------------

?s s1 and s2 and s3

409	S1
-----	----

4026	S2
------	----

40099	S3
-------	----

S4	1	S1 AND S2 AND S3
----	---	------------------

?t s4/3,k/all

**4/3,K/1 (Item 1 from file: 5)**

DIALOG(R) File 5:Biosis Previews(R)

(c) 2000 BIOSIS. All rts. reserv.

12137256 BIOSIS NO.: 199900432105

**Creation and characterization of \*transgenic\* \*mice\* expressing Cre recombinase in skeletal muscle and neurons.**

AUTHOR: Tiziano Francesco(a); Miniou P(a); Frugier T(a); Roblot N(a);

Dierich A(a); le Meur M(a); Melki J(a)

AUTHOR ADDRESS: (a)IGBMC, Illkirch, Strasbourg\*\*France

1999

JOURNAL: European Journal of Human Genetics 7 (SUPPL. 1):p113-114 July, 1999

CONFERENCE/MEETING: 31st Annual Meeting of the European Society of Human Genetics Geneva, Switzerland May 29-June 1, 1999

ISSN: 1018-4813

RECORD TYPE: Citation

LANGUAGE: English

**Creation and characterization of \*transgenic\* \*mice\* expressing Cre recombinase in skeletal muscle and neurons.**

DESCRIPTORS:

DISEASES: \*spinal\* \*muscular\* \*atrophy\*--

CHEMICALS & BIOCHEMICALS: ...murine \*SMN\* \*gene\* (Muridae...

?ds

Set	Items	Description
-----	-------	-------------

S1	409	(SMN (W) GENE?)
----	-----	-----------------

S2	4026	(SPINAL (W) MUSCULAR (W) ATROPHY)
----	------	-----------------------------------

S3	40099	(TRANSGENIC (W) (MOUSE OR MICE))
----	-------	----------------------------------

S4 1 S1 AND S2 AND S3  
 ?s s1 and s3  
 409 S1  
 40099 S3  
 S5 1 S1 AND S3  
 ?t s5/3,k/all

5/3,K/1 (Item 1 from file: 5)  
 DIALOG(R)File 5:Biosis Previews(R)  
 (c) 2000 BIOSIS. All rts. reserv.

12137256 BIOSIS NO.: 199900432105

**Creation and characterization of \*transgenic\* \*mice\* expressing Cre recombinase in skeletal muscle and neurons.**

AUTHOR: Tiziano Francesco(a); Miniou P(a); Frugier T(a); Roblot N(a); Dierich A(a); le Meur M(a); Melki J(a)

AUTHOR ADDRESS: (a)IGBMC, Illkirch, Strasbourg\*\*France  
 1999

JOURNAL: European Journal of Human Genetics 7 (SUPPL. 1):p113-114 July, 1999

CONFERENCE/MEETING: 31st Annual Meeting of the European Society of Human Genetics Geneva, Switzerland May 29-June 1, 1999

ISSN: 1018-4813

RECORD TYPE: Citation

LANGUAGE: English

**Creation and characterization of \*transgenic\* \*mice\* expressing Cre recombinase in skeletal muscle and neurons.**

DESCRIPTORS:

CHEMICALS & BIOCHEMICALS: ...murine \*SMN\* \*gene\* (Muridae...  
 ?ds

Set	Items	Description
S1	409	(SMN (W) GENE?)
S2	4026	(SPINAL (W) MUSCULAR (W) ATROPHY)
S3	40099	(TRANSGENIC (W) (MOUSE OR MICE))
S4	1	S1 AND S2 AND S3
S5	1	S1 AND S3

?s s2 and s3

	4026	S2
	40099	S3
S6	21	S2 AND S3

?rd

...completed examining records

S7	16	RD (unique items)
----	----	-------------------

?s s7 and (human (w) SMN (w) gene)

Processing

	16	S7
	15705149	HUMAN
	991	SMN
	1496029	GENE
	40	HUMAN (W) SMN (W) GENE
S8	0	S7 AND (HUMAN (W) SMN (W) GENE)

?s s7 and (knockout (w) mutation)

	16	S7
	19660	KNOCKOUT
	402502	MUTATION
	305	KNOCKOUT (W) MUTATION
S9	0	S7 AND (KNOCKOUT (W) MUTATION)

?s s1 and (transgenic (w) animal?)

Processing

	409	S1
	76050	TRANSGENIC
	14521177	ANIMAL?
	6613	TRANSGENIC (W) ANIMAL?
S10	0	S1 AND (TRANSGENIC (W) ANIMAL?)

?s s1 and (transgenic (w) rat?)

Processing

409 S1  
76050 TRANSGENIC  
6228380 RAT?  
1427 TRANSGENIC(W)RAT?  
S11 0 S1 AND (TRANSGENIC (W) RAT?)

?t s7/3,k/all

7/3,K/1 (Item 1 from file: 155)

DIALOG(R) File 155:MEDLINE(R)

(c) format only 2000 Dialog Corporation. All rts. reserv.

10367554 20122241

**The human centromeric survival motor neuron gene (SMN2) rescues embryonic lethality in Smn(-/-) mice and results in a mouse with \*spinal\* \*muscular\* \*atrophy\*.**

Monani UR; Sendtner M; Coover DD; Parsons DW; Andreassi C; Le TT; Jablonka S; Schrank B; Rossol W; Prior TW; Morris GE; Burghes AH  
Departments of Neurology, College of Medicine, Ohio State University, Columbus, OH 43210, USA.

Human molecular genetics (ENGLAND) Feb 12 2000, 9 (3) p333-9, ISSN 0964-6906 Journal Code: BRC

Contract/Grant No.: NS38650, NS, NINDS

Languages: ENGLISH

Document type: JOURNAL ARTICLE

**The human centromeric survival motor neuron gene (SMN2) rescues embryonic lethality in Smn(-/-) mice and results in a mouse with \*spinal\* \*muscular\* \*atrophy\*.**

Proximal \*spinal\* \*muscular\* \*atrophy\* (SMA) is a common motor neuron disease in humans and in its most severe form causes death by the age of 2 years. It is...

... SMN2 ). Mice possess only one survival motor neuron gene ( Smn ) whose loss is embryonic lethal. Therefore, to obtain a mouse model of SMA we created \*transgenic\* \*mice\* that express human SMN2 and mated these onto the null Smn (-/-) background. We show that Smn (-/-); SMN2 mice carrying one or two copies of the...

Chemical Name: Nerve Tissue Proteins; (SMN protein (\*spinal\* \*muscular\* \*atrophy\*))

7/3,K/2 (Item 2 from file: 155)

DIALOG(R) File 155:MEDLINE(R)

(c) format only 2000 Dialog Corporation. All rts. reserv.

10264509 20082811

**A mouse model for \*spinal\* \*muscular\* \*atrophy\*.**

Hsieh-Li HM; Chang JG; Jong YJ; Wu MH; Wang NM; Tsai CH; Li H

Institute of Molecular Biology, Academia Sinica, Taipei, Taiwan.

Nature genetics (UNITED STATES) Jan 2000, 24 (1) p66-70, ISSN 1061-4036 Journal Code: BRO

Languages: ENGLISH

Document type: JOURNAL ARTICLE

**A mouse model for \*spinal\* \*muscular\* \*atrophy\*.**

... motor neuron gene is present in humans in a telomeric copy, SMN1, and several centromeric copies, SMN2. Homozygous mutation of SMN1 is associated with proximal \*spinal\* \*muscular\* \*atrophy\* (SMA), a severe motor neuron disease characterized by early childhood onset of progressive muscle weakness. To understand the functional role of SMN1 in SMA, we produced mouse lines deficient for mouse Smn and \*transgenic\* \*mouse\* lines that expressed human SMN2. Smn-/- mice died during the peri-implantation stage. In contrast, \*transgenic\* \*mice\* harbouring SMN2 in the Smn-/- background showed pathological changes in the spinal cord and skeletal muscles similar to those of SMA patients. The severity of...



Chemical Name: DNA Primers; (Nerve Tissue Proteins; (SMN protein ( \*spinal\* \*muscular\* \*atrophy\*)

7/3,K/3 (Item 3 from file: 155)  
DIALOG(R) File 155:MEDLINE(R)  
(c) format only 2000 Dialog Corporation. All rts. reserv.

08567047 97173353

**Animal models of ALS.**  
Pioro EP; Mitsumoto H  
Cleveland Clinic Foundation, OH 44195, USA.  
Clinical neuroscience (UNITED STATES) 96 1995, 3 (6) p375-85, ISSN  
1065-6766 Journal Code: B9U  
Languages: ENGLISH  
Document type: JOURNAL ARTICLE; REVIEW; REVIEW, TUTORIAL

... models have been most extensively studied, including three mouse models: motor neuron degeneration (Mnd), progressive motor neuronopathy (pmn), wobbler, and one canine model: hereditary canine \*spinal\* \*muscular\* \*atrophy\* (HCSMA). The wobbler mouse has been the most extensively studied of these models with analyses of clinical, pathological (perikaryon, axon, muscle), and biochemical features. Experimentally...

... mouse models in which genes relevant to the human disease or motor neuron biology have been manipulated. The most clinically relevant of these is a \*transgenic\* \*mouse\* overexpressing the mutated SOD1 gene of FALS patients, which has already provided significant insights into mechanisms of motor neuron degeneration in this disease. Because no...

7/3,K/4 (Item 1 from file: 5)  
DIALOG(R) File 5:Biosis Previews(R)  
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12214817 BIOSIS NO.: 199900509666

**Creation and characterization of \*transgenic\* \*mouse\* lines expressing Cre recombinase in skeletal muscle or neurons.**

AUTHOR: Tiziano F(a); Miniou P(a); Frugier T(a); Dierich A(a); Le Meur M(a); Melki J(a)

AUTHOR-ADDRESS: (a)IGBMC. INSERM/CNRS/ULP, Illkirch, Strasbourg\*\*France  
1999

JOURNAL: American Journal of Human Genetics 65 (4):pA493 Oct., 1999

CONFERENCE/MEETING: 49th Annual Meeting of the American Society of Human Genetics San Francisco, California, USA October 19-23, 1999

SPONSOR: The American Society of Human Genetics

ISSN: 0002-9297

RECORD TYPE: Citation

LANGUAGE: English

**Creation and characterization of \*transgenic\* \*mouse\* lines expressing Cre recombinase in skeletal muscle or neurons.**

DESCRIPTORS:

DISEASES: \*spinal\* \*muscular\* \*atrophy\*--

7/3,K/5 (Item 2 from file: 5)  
DIALOG(R) File 5:Biosis Previews(R)  
(c) 2000 BIOSIS. All rts. reserv.

12137256 BIOSIS NO.: 199900432105

**Creation and characterization of \*transgenic\* \*mice\* expressing Cre recombinase in skeletal muscle and neurons.**

AUTHOR: Tiziano Francesco(a); Miniou P(a); Frugier T(a); Roblot N(a); Dierich A(a); le Meur M(a); Melki J(a)

AUTHOR ADDRESS: (a)IGBMC, Illkirch, Strasbourg\*\*France

1999

JOURNAL: European Journal of Human Genetics 7 (SUPPL. 1):p113-114 July, 1999

CONFERENCE/MEETING: 31st Annual Meeting of the European Society of Human Genetics Geneva, Switzerland May 29-June 1, 1999

ISSN: 1018-4813

RECORD TYPE: Citation

LANGUAGE: English

**Creation and characterization of \*transgenic\* \*mice\* expressing Cre recombinase in skeletal muscle and neurons.**

DESCRIPTORS:

DISEASES: \*spinal\* \*muscular\* \*atrophy\*--

7/3,K/6 (Item 1 from file: 73)

DIALOG(R)File 73:EMBASE

(c) 2000 Elsevier Science B.V. All rts. reserv.

07515254 EMBASE No: 1998422152

**The genetic and molecular mechanisms of motor neuron disease**

Wong P.C.; Rothstein J.D.; Price D.L.

P.C. Wong, Department of Pathology, The Johns Hopkins University, School of Medicine, 720 Rutland Avenue, Baltimore, MD 21205-2196 United States

AUTHOR EMAIL: pcwong@welchlink.welch.jhu.edu

Current Opinion in Neurobiology ( CURR. OPIN. NEUROBIOL. ) (United Kingdom) 1998, 8/6 (791-799)

CODEN: COPUE ISSN: 0959-4388

DOCUMENT TYPE: Journal; Review

LANGUAGE: ENGLISH SUMMARY LANGUAGE: ENGLISH

NUMBER OF REFERENCES: 84

MEDICAL DESCRIPTORS:

gene isolation; \*transgenic\* \*mouse\*; gene targeting; clinical feature; spastic paraplegia--congenital disorder--cn; \*spinal\* \*muscular\* \*atrophy\*--congenital disorder--cn; human; review; priority journal

7/3,K/7 (Item 2 from file: 73)

DIALOG(R)File 73:EMBASE

(c) 2000 Elsevier Science B.V. All rts. reserv.

07477051 EMBASE No: 1998395122

**Pathogenesis of neurodegenerative diseases associated with expanded glutamine repeats: New answers, new questions**

Ross C.A.; Margolis R.L.; Becher M.W.; Wood J.D.; Engelender S.; Cooper J.K.; Sharp A.H.

C.A. Ross, Johns Hopkins University, School of Medicine, Department of Psychiatry, 720 Rutland Avenue, Baltimore, MD 21205-2196 United States

AUTHOR EMAIL: caross@jhu.edu

Progress in Brain Research ( PROG. BRAIN RES. ) (Netherlands) 1998, 117/- (397-419)

CODEN: PBRRA ISSN: 0079-6123

DOCUMENT TYPE: Journal; Conference Paper

LANGUAGE: ENGLISH SUMMARY LANGUAGE: ENGLISH

NUMBER OF REFERENCES: 155

...polar zipper', into beta pleated sheets. Recent findings have now established the presence of such aggregates in selected regions of brain from affected individuals, in \*transgenic\* \*mice\* expressing expanded repeats, and in isolated cells transfected with expanded repeats. The aggregates are most prominently manifest as neuronal intranuclear inclusion bodies. As the investigation...

MEDICAL DESCRIPTORS:

\*huntington chorea--etiology--et; \*hereditary \*spinal\* \*muscular\* \*atrophy\*--etiology--et; \*trinucleotide repeat

neurologic disease--etiology--et; degenerative disease--etiology--et;  
\*spinal\* \*muscular\* \*atrophy\*--etiology--et; \*transgenic\* \*mouse\*; gene  
deletion; gene mutation; genetic predisposition; neurotoxicity--etiology  
--et; human; nonhuman; mouse; animal model; controlled study; human tissue;  
animal tissue; conference paper; priority journal

7/3,K/8 (Item 3 from file: 73)  
DIALOG(R)File 73:EMBASE  
(c) 2000 Elsevier Science B.V. All rts. reserv.

07273542 EMBASE No: 1998185616

**Androgen receptor YAC \*transgenic\* \*mice\* carrying CAG 45 alleles show trinucleotide repeat instability**

La Spada A.R.; Peterson K.R.; Meadows S.A.; McClain M.E.; Jeng G.;  
Chmelar R.S.; Haugen H.A.; Chen K.; Singer M.J.; Moore D.; Trask B.J.;  
Fischbeck K.H.; Clegg C.H.; McKnight G.S.

A.R. La Spada, Box 357110, University Washington Medical Center, 1959 NE  
Pacific Street, Seattle, WA 98195-7110 United States

AUTHOR EMAIL: laspada@mail.labmed.washington.edu

Human Molecular Genetics ( HUM. MOL. GENET. ) (United Kingdom) 1998, 7/6  
(959-967)

CODEN: HMGEE ISSN: 0964-6906

DOCUMENT TYPE: Journal; Article

LANGUAGE: ENGLISH SUMMARY LANGUAGE: ENGLISH

NUMBER OF REFERENCES: 42

**Androgen receptor YAC \*transgenic\* \*mice\* carrying CAG 45 alleles show trinucleotide repeat instability**

...alleles (37-66 CAGs) change in length when transmitted from parents to offspring, with a significantly greater tendency to shift size when inherited paternally. As \*transgenic\* \*mice\* carrying human AR cDNAs with 45 and 66 CAG repeats do not display repeat instability, we attempted to model trinucleotide repeat instability by generating \*transgenic\* \*mice\* with yeast artificial chromosomes (YACs) carrying AR CAG repeat expansions in their genomic context. Studies of independent lines of AR YAC \*transgenic\* \*mice\* with CAG 45 alleles reveal intergenerational instability at an overall rate of ~ 10%. We also find that the 45 CAG repeat tracts are significantly more unstable with maternal transmission and as the transmitting mother ages. Of all the CAG/CTG repeat \*transgenic\* \*mice\* produced to date the AR YAC CAG 45 mice are unstable with the smallest trinucleotide repeat mutations, suggesting that the length threshold for repeat instability...

**MEDICAL DESCRIPTORS:**

\*trinucleotide repeat; \*hereditary \*spinal\* \*muscular\* \*atrophy\*  
--congenital disorder--cn

yeast artificial chromosome; allele; X chromosome linkage; exon;  
\*transgenic\* \*mouse\*; dna flanking region; dna sequence; sequence tagged  
site; gene mapping; gene locus; polymerase chain reaction; parental age;  
immunoblotting; fluorescence in situ hybridization; nonhuman; mouse; animal  
...

7/3,K/9 (Item 4 from file: 73)  
DIALOG(R)File 73:EMBASE  
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07247164 EMBASE No: 1998136081

**Evidence of mdx mouse skeletal muscle fragility in vivo by eccentric running exercise**

Vilquin J.-T.; Brussee V.; Asselin I.; Kinoshita I.; Gingras M.; Tremblay J.P.

Dr. J.P. Tremblay, Centre de Recherche en Neurobiologie, Univ. Laval and  
Hopital Enfant-Jesus, 1401, 18e Rue, Laval, Que. G1J 1Z4 Canada  
Muscle and Nerve ( MUSCLE NERVE ) (United States) 1998, 21/5 (567-576)

CODEN: MUNED ISSN: 0148-639X  
DOCUMENT TYPE: Journal; Article  
LANGUAGE: ENGLISH SUMMARY LANGUAGE: ENGLISH  
NUMBER OF REFERENCES: 51

...of 18 months, and the role of dystrophin in fiber integrity is not fully understood. The fragility of the skeletal muscle fibers was investigated in \*transgenic\* \*mice\* expressing beta-galactosidase under the control of a muscle specific promoter. Adult mdx/beta-galactosidase (dystrophin-negative) and normal/beta-galactosidase (dystrophin-positive) mice were submitted...

**MEDICAL DESCRIPTORS:**

running; exercise; hereditary \*spinal\* \*muscular\* \*atrophy\*--diagnosis--di; hereditary \*spinal\* \*muscular\* \*atrophy\*--etiology--et; enzyme analysis; protein expression; phenotype; nonhuman; male; female; mouse; animal model; controlled study; animal tissue; animal cell; article; priority journal

7/3,K/10 (Item 5 from file: 73)  
DIALOG(R) File 73:EMBASE  
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07084073 EMBASE No: 1997365936

**\*Transgenic\* \*mouse\* models of neurodegenerative disease caused by CAG/polyglutamine expansions**

Bates G.P.; Davies S.W.

Dr. G.P. Bates, Medical and Molecular Genetics, UMDS, Guy's Hospital, London SE1 9RT United Kingdom

AUTHOR EMAIL: g.bates@umds.ac.uk

Molecular Medicine Today ( MOL. MED. TODAY ) (United Kingdom) 1997, 3/11 (508-518)

CODEN: MMTOF ISSN: 1357-4310

PUBLISHER ITEM IDENTIFIER: S1357431097011428

DOCUMENT TYPE: Journal; Review

LANGUAGE: ENGLISH SUMMARY LANGUAGE: ENGLISH

NUMBER OF REFERENCES: 47

**\*Transgenic\* \*mouse\* models of neurodegenerative disease caused by CAG/polyglutamine expansions**

**MEDICAL DESCRIPTORS:**

animal model; cell death; gene expression; gene mutation; genetic stability; huntington chorea--etiology--et; machado joseph disease--etiology--et; molecular genetics; mouse; nonhuman; phenotype; review; \*spinal\* \*muscular\* \*atrophy\*--etiology--et; spinocerebellar degeneration--etiology--et; \*transgenic\* \*mouse\*

7/3,K/11 (Item 6 from file: 73)  
DIALOG(R) File 73:EMBASE  
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06965185 EMBASE No: 1997249782

**The CAG/polyglutamine tract diseases: Gene products and molecular pathogenesis**

Koshy B.T.; Zoghbi H.Y.

Dr. B.T. Koshy, Howard Hughes Medical Institute, Baylor College of Medicine, One Baylor Plaza, Houston, TX 77030 United States

Brain Pathology ( BRAIN PATHOL. ) (United States) 1997, 7/3 (927-942)

CODEN: BRPAE ISSN: 1015-6305

DOCUMENT TYPE: Journal; Conference Paper

LANGUAGE: ENGLISH SUMMARY LANGUAGE: ENGLISH

NUMBER OF REFERENCES: 157

...polyglutamine confers a gain of function onto the involved protein. To understand the mechanisms underlying the pathogenesis of these diseases, investigators have turned to generating \*transgenic\* \*mice\* which

recapitulate some of the features of the human disease and hence are excellent model systems to study the progression of the disease in vivo.

**MEDICAL DESCRIPTORS:**

\*fragile X syndrome--etiology--et; \*\*spinal\* \*muscular\* \*atrophy\*--etiology--et; \*spinocerebellar degeneration--etiology--et

**7/3,K/12 (Item 7 from file: 73)**

DIALOG(R)File 73:EMBASE

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06725751 EMBASE No: 1997007213

**Glutamine repeats and inherited neurodegenerative diseases: Molecular aspects**

Perutz M.F.

M.F. Perutz, MRC Laboratory of Molecular Biology, Hills Road, Cambridge CB2 2QH United Kingdom

Current Opinion in Structural Biology ( CURR. OPIN. STRUCT. BIOL. ) ( United Kingdom) 1996, 6/6 (848-858)

CODEN: COSBE ISSN: 0959-440X

DOCUMENT TYPE: Journal; Review

LANGUAGE: ENGLISH SUMMARY LANGUAGE: ENGLISH

NUMBER OF REFERENCES: 62

...41 repeats. Protein constructs with more than 41 repeats are toxic to E. coli and to CHO cells in culture, and they elicit ataxia in \*transgenic\* \*mice\*. These observations argue in favour of a distinct change of structure associated with elongation beyond 37-41 glutamine repeats. The review describes experiments designed to...

**MEDICAL DESCRIPTORS:**

ataxia; cho cell; cytotoxicity; dentatorubropallidoluysian atrophy--etiology--et; dentatorubropallidoluysian atrophy--congenital disorder--cn; dna sequence; escherichia coli; hereditary \*spinal\* \*muscular\* \*atrophy\*--etiology--et; hereditary \*spinal\* \*muscular\* \*atrophy\*--congenital disorder--cn; human; huntington chorea--congenital disorder--cn; huntington chorea--etiology--et; hydrogen bond; machado joseph disease--congenital disorder--cn; machado joseph disease--etiology--et; mouse; nonhuman; oligomerization; priority journal; protein conformation; protein folding; review; spinocerebellar degeneration--congenital disorder--cn; spinocerebellar degeneration--etiology--et; structure activity relation; \*transgenic\* \*mouse\*

**7/3,K/13 (Item 8 from file: 73)**

DIALOG(R)File 73:EMBASE

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06716854 EMBASE No: 1996381838

**Animal models for motor neuron diseases: Research directions**

Ludolph A.C.

Department of Neurology, University of Ulm, Steinhovelstrasse 9, 89075 Ulm Germany

Neurology ( NEUROLOGY ) (United States) 1996, 47/6 SUPPL. 4 (S228-S232)

CODEN: NEURA ISSN: 0028-3878

DOCUMENT TYPE: Journal; Review

LANGUAGE: ENGLISH SUMMARY LANGUAGE: ENGLISH

...neuron degenerative, wobbler, wasted, and autosomal-recessive progressive motor neuropathy mouse models), sporadically occurring (e.g., equine motor neuron disease), and experimentally induced (e.g., \*transgenic\* \*mice\* carrying a mutation in the gene encoding human Cu/Zn superoxide dismutase, toxic models) disorders. The models currently have three major drawbacks. First, there is...

**MEDICAL DESCRIPTORS:**

animal model; anterior horn cell disease--etiology--et; anterior horn cell disease--diagnosis--di; gene mutation; hereditary \*spinal\* \*muscular\*

\*atrophy\*--diagnosis--di; hereditary \*spinal\* \*muscular\* \*atrophy\*  
--etiology--et; lipofuscinosis--etiology--et; lipofuscinosis--diagnosis--di  
; nonhuman; priority journal; pyramidal tract; review; \*transgenic\* \*mouse\*

7/3,K/14 (Item 9 from file: 73)  
DIALOG(R)File 73:EMBASE  
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06529216 EMBASE No: 1996194816

**Transgenic and gene-targeting approaches to model disorders of motor neurons**

Wong P.C.; Borchelt D.R.; Lee M.K.; Pardo C.A.; Sisodia S.S.; Cleveland D.W.; Koliatsos V.E.; Price D.K.  
Department of Pathology, Johns Hopkins Univ. School Medicine, Baltimore, MD United States  
Seminars in the Neurosciences ( SEMIN. NEUROSCI. ) (United Kingdom) 1996, 8/3 (163-169)  
CODEN: SNEUE ISSN: 1044-5765  
DOCUMENT TYPE: Journal; Article  
LANGUAGE: ENGLISH SUMMARY LANGUAGE: ENGLISH

...Over the past several years, significant progress has been made in understanding the genetics of some of these disorders, including familial amyotrophic lateral sclerosis (FALS), \*spinal\* \*muscular\* \*atrophy\* (SMA), and spinal bulbar muscular atrophy (SBMA). For example, some of the autosomal dominant cases of FALS are linked to mutations in the superoxide dismutase 1 (SOD1) gene. Several groups have introduced these SOD1 mutations into \*transgenic\* \*mice\*, and these animals develop features of the human disease. Other investigators have used transgenic strategies to overexpress wild-type (wt) or mutant neurofilament (NF) genes...

**MEDICAL DESCRIPTORS:**

...sclerosis--etiology--et; animal experiment; animal model; article; cell death; cell inclusion; cytoskeleton; gene mutation; hyperreflexia; mouse; muscle atrophy; muscle weakness; neurofilament; nonhuman; phenotype; spasticity; \*spinal\* \*muscular\* \*atrophy\*--etiology--et; \*transgenic\* \*mouse\*

7/3,K/15 (Item 10 from file: 73)  
DIALOG(R)File 73:EMBASE  
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06217067 EMBASE No: 1995245815

**Genetic approaches to pathogenesis of neurodegenerative diseases**

Orr H.T.; Clark H.B.  
Laboratory Medicine/Pathology Dept., Institute of Human Genetics, University of Minnesota, Minneapolis, MN United States  
Laboratory Investigation ( LAB. INVEST. ) (United States) 1995, 73/2 (161-171)  
CODEN: LAINA ISSN: 0023-6837  
DOCUMENT TYPE: Journal; Review  
LANGUAGE: ENGLISH

**MEDICAL DESCRIPTORS:**

...etiology--et; machado joseph disease--etiology--et; major clinical study; male; nerve cell necrosis; neurologic disease--etiology--et; neuropathology; nonhuman; onset age; priority journal; review; \*spinal\* \*muscular\* \*atrophy\*--etiology--et; \*transgenic\* \*mouse\*

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06019153 EMBASE No: 1995049283

**Stability of an expanded trinucleotide repeat in the androgen receptor gene in \*transgenic\* \*mice\***

Bingham P.M.; Scott M.O.; Wang S.; McPhaul M.J.; Wilson E.M.; Garbern J.Y.; Merry D.E.; Fischbeck K.H.  
Division of Neurology, Children's Hospital of Philadelphia, Philadelphia, PA 19104 United States  
Nature Genetics ( NAT. GENET. ) (United States) 1995, 9/2 (191-196)  
CODEN: NGENE ISSN: 1061-4036  
DOCUMENT TYPE: Journal; Article  
LANGUAGE: ENGLISH SUMMARY LANGUAGE: ENGLISH

**Stability of an expanded trinucleotide repeat in the androgen receptor gene in \*transgenic\* \*mice\***

...stability of a trinucleotide repeat and to develop an animal model of one of these disorders, spinal and bulbar muscular atrophy (SBMA), we have generated \*transgenic\* \*mice\* carrying either the normal or expanded repeat human androgen receptor (AR) gene. Unlike the disease allele in humans, the AR cDNA containing the expanded repeat in \*transgenic\* \*mice\* showed no change in repeat length with transmission. Expression of the SBMA AR was found in \*transgenic\* \*mice\*, but at a lower level than normal endogenous expression. The lack of a physiological pattern of expression may explain why no phenotypic effects of the...

**MEDICAL DESCRIPTORS:**

\*\*spinal\* \*muscular\* \*atrophy\*; \*transgene  
animal model; animal tissue; article; controlled study; densitometry; gene expression; mouse; nonhuman; phenotype; polyacrylamide gel electrophoresis; polymerase chain reaction; priority journal; southern blotting;  
\*transgenic\* \*mouse\*  
?ds

Set	Items	Description
S1	409	(SMN (W) GENE?)
S2	4026	(SPINAL (W) MUSCULAR (W) ATROPHY)
S3	40099	(TRANSGENIC (W) (MOUSE OR MICE))
S4	1	S1 AND S2 AND S3
S5	1	S1 AND S3
S6	21	S2 AND S3
S7	16	RD (unique items)
S8	0	S7 AND (HUMAN (W) SMN (W) GENE)
S9	0	S7 AND (KNOCKOUT (W) MUTATION)
S10	0	S1 AND (TRANSGENIC (W) ANIMAL?)
S11	0	S1 AND (TRANSGENIC (W) RAT?)

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PMID: 10825724

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Hum Mol Genet. 2000 Mar 22;9(5):849-58.

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Exclusion of the beta-subunit of type II calmodulin kinase for the wobbler spinal muscular atrophy gene.

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The 21-aminosteroid U-74389F increases the number of glial fibrillary acidic protein-expressing astrocytes in the spinal cord of control and Wobbler mice.

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Genomics. 1995 Sep 1;29(1):131-5.

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Related Article

Stability of an expanded trinucleotide repeat in the androgen receptor gene in transgenic mice.

Nat Genet. 1995 Feb;9(2):191-6.

PMID: 7719348; UI: 95235566

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PMID: 7416238; UI: 81019238

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